

# **Surgical Correction of Chest Wall Deformities**

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# **Background**

Chest wall deformities result from abnormal growth of the rib cartilages which pushes the sternum either inward or outward, away from the plane of the chest. The deformities can range from mild, symmetric indentions or protrusions, to severe asymmetric deformities. The appearance of the deformity often changes dramatically around the time of adolescent growth. Chest wall deformities may be corrected using various techniques; most require surgical intervention.

### **Definitions**

Pectus carinatum (PC)

Pectus carinatum (i.e., pigeon breast or chicken breast) is a congenital chest deformity characterized by an anterior protrusion deformity of the sternum and costal cartilages. PC is typically not confirmed until after the growth spurts of early adolescence. This deformity produces a rigid chest and, while symptoms are uncommon, it may result in inefficient respiration as a result of the restrictive chest formation. Three types of PC-related defects have been identified:

- Anterior displacement of the body of the sternum and symmetrical concavity of the costal cartilages
- Lateral depression of the ribs on one or both sides of the sternum
- Pouter pigeon breast (least common deformity) a defect that consists of an upper or chondromalacial prominence with protrusion of the manubrium and depression of the sternal body

The degree of physiological impairment is related to the degree of chest deformity. Patients with PC may develop symptoms as a result of restricted air exchange; complete expiration of air from the lungs may not occur. In addition, pain may result from the secondary pressures that develop from the overgrowth of cartilage. Other conditions that may be associated with PC include frequent respiratory infections, asthma, rickets, mitral valve disease, Marfan's syndrome, scoliosis and other cardiac changes.

# Pectus excavatum (PE)

Pectus excavatum is a posterior depression of the sternum and adjacent costal cartilages; often a cosmetic defect, but which may have varied anatomic and symptomatic presentations.

Surgical correction of pectus excavatum improves physical appearance in most patients and cardiorespiratory function in some, but the indications for intervention are not fully standardized. Standard PE surgical procedures; e.g.:

- Ravitch procedure standard open surgical technique that involves removing the ends of the ribs in the area that is depressed at the sternum. The sternum is then straightened out at the point it turns downward by breaking it horizontally. Stitches and a metal bar are used to hold the sternum in place under the skin. After two to three years, when remolding has taken place, the bar may be removed.
- Nuss procedure (aka minimally invasive repair of pectus excavatum [MIRPE]) closed procedure that corrects the pectus defect without cartilage resection by applying outward pressure to the sternum at the point of maximal inward deflection using a custom-contoured steel bar ("Nuss bar"). The Nuss bar is placed in the pleural space, passed behind the sternum, rotated 180 degrees, and then attached laterally to the outer edge of the rib cage. The bar is left in place for several months or years.

Investigational PE treatment approaches (see Limitations/Exclusions); e.g.:

- Sternal magnet (Magnetic Mini Mover procedure) designed to lift the sternum using magnetic attraction between a magnet attached to the sternum and another magnet on an external sternal brace.
- Sternal suction suction applied externally to the sternum to reduce sternal depression by about 1 cm per month. The suction device is used for one or more hours daily for 12 to 15 months. The device has also been used as an adjunct to conventional surgical correction.

# Poland Syndrome

Poland syndrome (i.e., Poland's anomaly, Poland's syndactyly) — rare congenital disorder associated with lateral depression of the ribs on one or both sides of the sternum. The right side of the body is affected twice as often as the left. When the anomaly occurs on the left side of the body, the heart and lungs are vulnerable, because they may be covered only by skin, fascia and pleura.

Although the anomaly is associated with a wide range of malformations, the condition is characterized by absence or hypoplasia of the pectoralis major muscle, absence or hypoplasia of the pectoralis minor muscle, absence of costal cartilages, hypoplasia of the breast and subcutaneous tissue, and a variety of hand and upper-extremity anomalies.

In cases of severe cartilage deficiency, patients may develop lung hernia and paradoxical respiratory motion. In less severe cases, patients may develop a simple flattening of the anterior chest wall.

Poland syndrome surgery techniques include, but may not be limited to: augmentation with tissue from the opposite breast, musculocutaneous flap to fill hollow space on the exterior of the chest, prosthetic augmentation, and surgical repair of the chest wall.

#### **Haller Index**

Aka pectus index (PI) or pectus severity index (PSI)

The Haller index, also called the pectus index (PI) or pectus severity index (PSI), is the most commonly used scale for determining the severity of chest wall deformities.

The index is defined as the width of the chest divided by the distance between the sternum and spine at the point of maximal depression.

The normal value is 2.54. In individuals with PE, a lower PSI indicates a more severe deformity in contrast to individuals with excavatum, in which a higher PSI indicates a more severe deformity. An index greater than 3.25 is considered severe for PE. Computerized tomography (CT) or magnetic resonance imaging (MRI) may be used to determine the index.

### **Related Medical Guidelines**

**Breast Implants and Reconstruction** 

**Cosmetic Surgery Procedures** 

### Guideline

Note: Coverage for the surgical repair of a chest wall deformity is dependent upon benefit plan language, may be subject to the provisions of a cosmetic and/or reconstructive surgery benefit and may be governed by state/federal mandates. Under many benefit plans, surgery for a chest wall deformity is not covered when performed solely for the purpose of improving or altering appearance or self-esteem or to treat psychological symptomatology or psychosocial complaints related to one's appearance. This includes, but is not limited to, treatments, drugs, products, hospital/facility charges, anesthesia, pathology/lab fees, radiology fees and professional fees by the surgeon, assistant surgeon, consultants and attending physicians

- Surgical procedures performed solely for cosmetic/psychological reasons are not considered medically necessary. (See <u>Limitations/Exclusions</u>)
- II. Surgical procedures performed to correct physiologic complications of Pectus Excavatum, Pectus Carinatum, or Poland syndrome are considered medically necessary and reconstructive when the following criteria are met:.
  - **A.** Functional impairment documented by one of the following:
    - Decreased cardiac output and/or abnormal pulmonary function during exercise
    - 2. Anticipation of future cardiovascular compromise
    - 3. Signs or symptoms that impair the member's ability to participate in usual activities (i.e., shortness of breath [dyspnea] at rest or on exertion)
    - 4. Arrhythmias or clinical stigmata of decreased cardiac output

AND

**B.** The procedure is expected to correct the functional impairment

AND

- **C.** The anatomical criterion for the condition is met:
  - 1. For treatment of PE, the Haller Index is  $\geq 3.25$
  - 2. For treatment of PC, the Haller Index is  $\leq 2.0$
  - 3. For treatment of Poland syndrome, when rib formation is absent

### **Limitations/Exclusions**

- **I.** EmblemHealth does not consider surgery for chest wall deformities to be medically necessary when performed for any of the following reasons:
  - **A.** Improve/alter appearance
  - B. Increase self-esteem
  - **C.** Treat psychological symptomatology or psychosocial complaints
- II. Bracing and surgical procedures to correct PC are considered cosmetic and not medically necessary when the deformity does not cause physiologic disturbances from compression of the heart or lungs.
- III. The following surgical procedures for PE are not considered medically necessary due to insufficient evidence of therapeutic value and are therefore not covered:

- A. Magnetic Mini Mover Procedure (3MP)
- B. Sternal suction (e.g., The Vacuum Bell)

# **Revision History**

Nov. 11, 2019 —removed dynamic compression bracing from Limitations/Exclusions

Oct. 12, 2018 — consolidated separate criteria sets for PE, PC and Poland syndrome into one criterion that emphasizes the presence of functional impairment and differentiated Haller Index parameters for PE and PC.

Nov. 11, 2016 — coverage limitations removed for Poland Syndrome.

# Medical Procedure and ICD-10 Diagnostic Coding

# **PECTUS EXCAVATUM**

### **Procedure Codes**

21740	Reconstructive repair of pectus excavatum or carinatum; open
21742	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), without thoracoscopy
21743	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), with thoracoscopy

# **ICD-10 Codes**

J44.9	Chronic obstructive pulmonary disease, unspecified
J98.4	Other disorders of lung
Q67.6	Pectus excavatum
R94.2	Abnormal results of pulmonary function studies

# PECTUS CARINATUM

# **Procedure Codes**

21740	Reconstructive repair of pectus excavatum or carinatum; open
21742	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), without thoracoscopy
21743	Reconstructive repair of pectus excavatum or carinatum; minimally invasive approach (Nuss procedure), with thoracoscopy

### **ICD-10 Codes**

Q67.7	
	Pectus carinatum

### **POLAND SYNDROME**

#### **Procedure Codes**

15734	Muscle, myocutaneous, or fasciocutaneous flap; trunk
15756	Free muscle or myocutaneous flap with microvascular anastomosis
20900	Bone graft, any donor area; minor or small (e.g., dowel or button)
20902	Bone graft, any donor area; major or large

### **ICD-10 Codes**

Q79.8	Other congenital malformations of musculoskeletal system
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